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Beware of the persistent abscess: Chronic granulomatous disease presenting with suppurative lymphadenitis

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ABSTRACT

An 18-month old boy presented with axillary suppurative lymphadenitis, which failed to respond to conservative management. His care was transferred to pediatric surgeons for further management. Resolution of the abscess was not achieved despite two incision and drainage procedures. Cultures obtained from the abscess grew *Serratia Marcescens*, and *Staphylococcus aureus*. A diagnosis of chronic granulomatous disease (CGD) was made, subsequently identified to be x-linked. Review of a chest X-ray previously reported as normal identified focal consolidation. This was investigated with CT and thought to represent previous granuloma with current *Staphylococcus* infection. The boy was started on prophylactic septrin and itraconizole, in addition to 2 months of treatment antibiotics. The abscess healed, and a repeat CT demonstrated resolution of the cavitating lung lesion. A gastrostomy was inserted in preparation for a T cell-depleted haploidentical bone marrow transplantation, which to date appears to have been curative. Chronic granulomatous disease is a rare genetic primary immunodeficiency associated with high mortality if diagnosis is delayed. The recent establishment of a UK CGD registry has identified a far higher prevalence than previously thought. Stem cell transplantation now offers a potentially curative treatment, with better outcomes in younger children. It is therefore essential that pediatric surgeons are aware of this diagnosis, and that appropriate investigations are instituted when an abscess fails to resolve, or has cultured unusual organisms.

1. Case report

An 18 month-old boy was transferred from his local hospital for further management of right axillary suppurative lymphadenitis. One week prior to this, a lesion with a punctum on his right chest wall had been noted. This had discharged spontaneously and scabbed over. The axillary lymphadenitis had been treated with a 4-day course of intravenous benzyl penicillin and flucloxacillin with no improvement. An ultrasound showed features of a thick walled abscess. The child was systemically well, with a white cell and neutrophil count within the normal range, but a raised c-reactive protein at 26.

An incision and drainage was performed, during which a small amount of pus was obtained and sent for microscopy and culture. The boy was discharged home without antibiotics.

Over the next 3 days, the abscess was noted to re-accumulate, and a new lesion developed on the left chest wall. A course of oral co-amoxiclav was commenced. The boy was reviewed by the pediatric surgery team who noted the swelling had reduced, but frank pus was draining from the wound. Cultures were obtained from the new chest wall lesion and the original abscess cavity, and sent for microbiology and virology. The swab from the abscess grew *Staphylococcus aureus*, resistant to penicillin, but sensitive to flucloxacillin, which was then started. The swab from the new chest wall lesion was negative for herpes and varicella.

Three days later, the boy was re-admitted as the axillary abscess showed no evidence of resolution, with copious discharge of pus. Intravenous flucloxacillin was commenced, and further swabs were sent to exclude atypical mycobacteria. The boy was reviewed by a pediatric infectious disease specialist. There was no history of recent travel, no contact with TB and no past medical history of note – specifically no previous boils, abscesses or recurrent infections. The rest of the family were fit and well. On examination there was no cervical, inguinal or left axillary lymphadenopathy, and no hepatosplenomegaly. The impression was that these lesions were secondary to a virulent *Staphylococcus aureus* infection, and panton valentine leukocidin (PVL) testing was requested. A chest X-ray was performed to exclude pulmonary lesions and underlying

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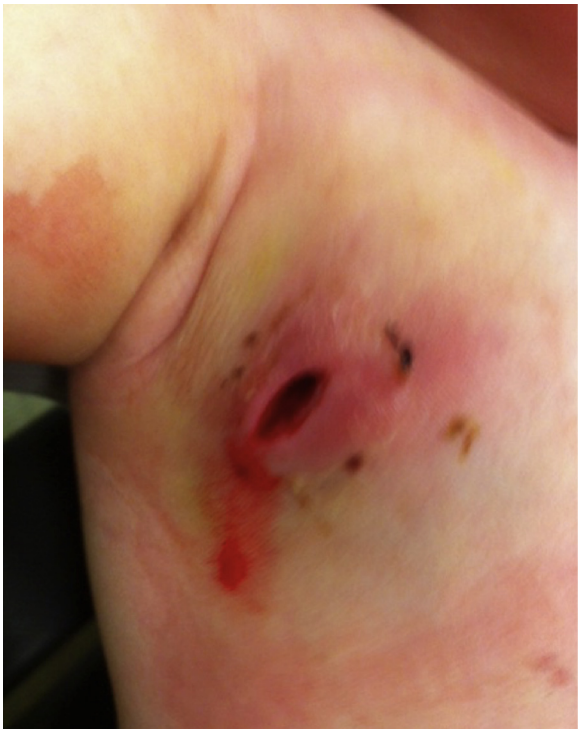

Fig. 1. Right axillary abscess following 2nd incision and drainage.

osteomyelitis of the right lateral ribs, and was reported as normal. A further incision and drainage was performed and a 10-day course of clindamycin (initially intravenous, then oral) was added to the flucloxacillin (Fig. 1).

The delayed culture result from the first incision and drainage revealed growth of *Serratia marcescens*. As the boy was well, his clindamycin was stopped and he continued flucloxacillin orally. When the culture result from the second incision and drainage also revealed growth of *Serratia marcescens*, oral ciprofloxacin was added to flucloxacillin. He was discharged home to complete a 14-day course of antibiotics.

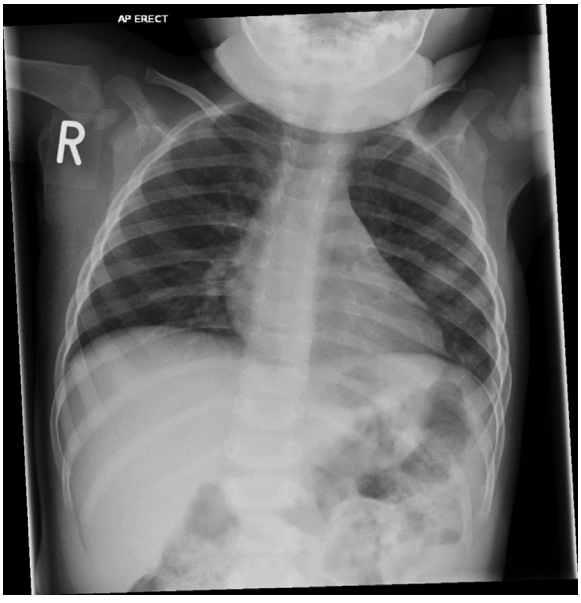

Fig. 2. Chest X-ray demonstrating left mid zone consolidation.

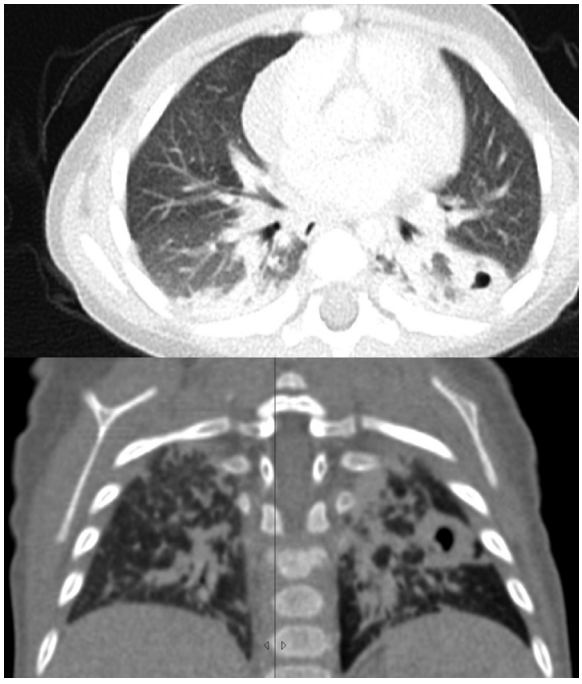

Fig. 3. CT chest revealed cavitating lesion in left lower lobe thought to represent previous granuloma and current *Staphylococcus aureus* infection.

At the first dressing change 2 days post-discharge, there was persistent offensive pus. As the cultures from the abscess had grown *Serratia marcescens* and *Staphylococcus aureus*, concern was raised regarding a possible diagnosis of chronic granulomatous disease (CGD). On the advice of the pediatric infectious disease specialist, bloods were sent for a nitroblue tetrazolium (NBT) assay, lymphocyte subsets, immunoglobulins and vaccine response. The NBT assay confirmed a diagnosis of CGD.

A PICC line was inserted and he was started on intravenous meropenem and prophylactic itraconazole. Liver and splenic abscesses were excluded with an ultrasound. Review of the initial chest X-ray raised concern regarding left mid zone focal consolidation. CT chest revealed a cavitating focal area of consolidation within the left lower lobe, with multiple focal areas of calcification within the left hilum, mediastinum and left lung. A specialist opinion was sought, and it was felt that the appearances were suggestive of previous granuloma and current *Staphylococcus aureus* infection, with no typical features of fungal lung infection (Figs. 2 and 3). Aspergillus titres and T-spot were negative.

The CT chest was repeated following a 2-month course of antibiotics. The cavitating lesion had resolved completely, with persistence of chronic calcific changes. Intravenous meropenem was stopped, prophylactic septrin commenced and prophylactic itraconizole continued (Fig. 4).

The boy was confirmed to have x-linked CGD. A gastrostomy was inserted in preparation for a T cell-depleted haploidentical bone marrow transplant. He has made a very good recovery and now has essentially normal neutrophil function.

2. Discussion

Chronic granulomatous disease (CGD) is a rare genetic primary immunodeficiency, associated with a high mortality if diagnosis is delayed. Prevalence varies worldwide from 1 case per million to 1